INSTITUTE: National Institute of Arthritis and Musculoskeletal and Skin Diseases

STUDY NUMBER: 91-AR-0196 PRINCIPAL INVESTIGATOR: James D. Katz, M.D.

STUDY TITLE: Studies on the Natural History and Pathogenesis of Polymyositis, Dermatomyositis, and Related Diseases

Continuing Review Approved by the IRB on 01/06/15
Amendment Approved by the IRB on 07/29/15 (AA) Date Posted to Web: 07/31/15
Assent

You are being asked to participate in a study designed to learn more about the weakness of your muscles. Weakness can be caused by many things. The type of weakness we are looking for is the result of inflammation, or swelling, in the muscle. We are trying to learn more about what may cause the weakness and what medicines are best to use to treat it. Several doctors and other medical professionals will ask you questions and examine you.

You will be undergoing blood tests and other tests to measure how well your muscles and other parts of your body function (such as your lungs, heart, and mouth). The hazards of testing your blood primarily involve the pain of the needle puncturing the skin and the risk of getting a bruise. Very rarely, an infection may occur. You may also be asked to undergo some or all of the following tests.

HIV Testing at the NIH in Research

As part of this study, we may test you for infection with the human immunodeficiency virus (HIV), the virus that causes AIDS. If you are infected with HIV you will still be able to participate in this study. We will tell you what the results mean, how to find care, how to avoid infecting others, how we report HIV infection, and the importance of informing your partners at possible risk because of your HIV infection.

For those procedures with an asterisk (*) beside the number, you will be given a thorough explanation and asked to sign a separate consent form.

1*) Muscle biopsy. This procedure is performed in the treatment room on the ward. After a local anesthetic numbs the skin, a one-half inch to one-inch long incision is made in the thigh or upper arm, and a small piece of muscle is removed. The skin is closed with self-absorbing stitches.

Discomforts and Risks: Discomfort during the procedure will be avoided by the use of a local anesthetic. The biopsy site may be tender for several days. Risks include the rare complications of local bleeding or infection. A small scar will remain permanently at the biopsy site.

2*) Electromyography (EMG). This measures the electrical activity of the muscles. It involves the insertion of a needle into a muscle, usually through the skin, to record its electrical activity.

Discomforts and Risks: You may experience pain at the site of needle entry. There is a slight possibility of infection or bleeding.

3*) Magnetic resonance imaging (MRI). This is a new and sensitive diagnostic tool that uses a strong magnetic field and radio waves to show changes in tissue. There is no radiation risk. You will lie on a table in a space enclosed by a metal cylinder (the scanner itself). The time required to stay in the cylinder will be about 20-30 minutes. During part of the time, you will be asked to lie very still.

Discomforts and Risks: Patients are at risk for injury from MRI if they have metal objects in their bodies such as a pacemaker, aneurysm clips (metal clips on the wall of a large artery), artificial joints, inner ear implants, or shrapnel fragments. Welders and metal workers are also at risk for eye injury because of unsuspected tiny metal fragments there. Individuals with fear of confined spaces may become anxious during MRI. You will hear a thumping noise created by the radio waves forming the images. You will feel no pain, but you may find the noise and the closed-in space discomforting. You will be observed at all times by the operators and will be able to speak to them; you can be moved out of the machine at your request.

4) Standardized muscle testing by a physiotherapist, to see how strong you are;

5*) Swallowing studies with ultrasound and X-rays (barium swallow), to check your speaking and swallowing abilities;

6) Pulmonary function tests (PFT), to check your breathing, and, if necessary, chest X-rays and other tests;

7) Electrocardiogram (EKG), and, if necessary, other tests to measure your heart.

Other tests not listed may be recommended to help us plan the best treatment for you.

For all of the tests, you and your parents or guardian will be consulted in advance, given full information, and asked to approve them. If you or your parent or guardian does not approve of any of these tests, it will not be done. The results of the tests that we do, including the results of HIV testing or a pregnancy test done prior to any X-rays, will be discussed with you and, if you agree, with your parents or guardian.

Blood, muscle specimens, and any other material you donate may be used for laboratory research studies, possibly including DNA tests for genetic markers that correlate with your disease, and those specimens will be labeled with your name for reference purposes. On occasion, a coded serum specimen without your name on it may be sent to an outside laboratory for diagnostic tests. That coded specimen may also be tested for other antibodies for research purposes by that laboratory, but your name will not be sent to that laboratory. We will inform you and your physician of any genetic findings that would be helpful to you or your family, and will help to arrange for genetic counseling if that is needed. We plan no other genetic tests on that sample, and we will perform no other studies on it in our laboratory without obtaining your consent. We may provide a sample of the DNA to another laboratory, but only after removing your name and any marking that could identify you in any way**.**

Any tests necessary for your treatment will be done after separate discussions with you and your parent or guardian.

If you do not wish to have any of these tests or change your mind, the test will not be done.

I have had this study explained to me in a way that I understand, and I have had the chance to ask questions. I agree to take part in this study.

Signature of Minor Patient:_____ Date:_____

Print Name:_____

Signature of Investigator:_____ Date:_____

Print Name:_____